Asymptomatic Solitary Dermal Plaque

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Case Report

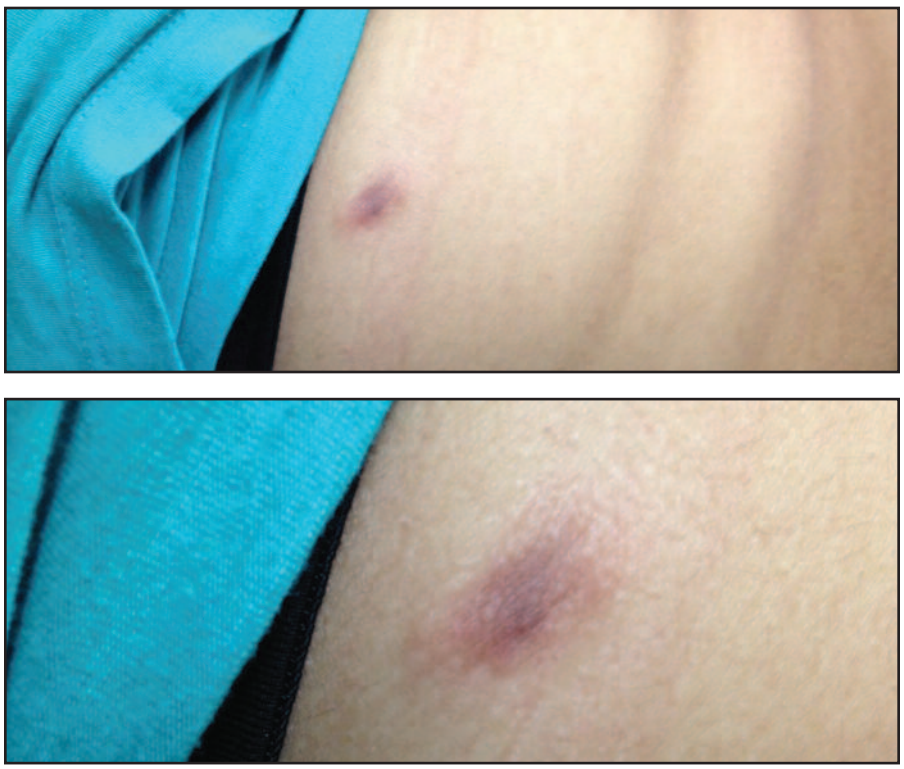
A 17-year-old Hispanic girl presented with a one-year history of a nonpruritic violaceous plaque on her right flank. The patient denied any antecedent trauma and there was no significant past medical history. Family history was notable only for a sister with mild plaque psoriasis. On physical examination, there was a 1.2cm firm, nontender, violaceous dermal plaque (Figures 1A and 1B). A 4mm punch biopsy was obtained from the center of the lesion (Figure 2) followed by a complete excision (Figures 3A and 3B).

Diagnosis

Cellular neurothekeoma (CNTK)

Microscopic Findings and Clinical Course

The biopsy specimen showed multiple nodules of epithelioid cells with pale cytoplasm and mild nuclear pleomorphism in the dermis (Figure 2). Immunohistochemical staining was positive for NK1/C3 and vimentin with weak positivity for CD68. Immunohistochemical staining was negative for S100, Melan-A, smooth muscle actin, desmin, and cytokeratin. An excision specimen revealed a deeper spindled component with a marked plexiform growth pattern in the subcutis that had features resembling a plexiform fibrohistiocytic tumor (PFHT) (Figures 3A and 3B). Although CNTKs are benign neoplasms that occasionally recur, PFHTs are considered borderline neoplasms that recur more frequently and rarely metastasize to regiona lymph nodes or to the lungs. For this reason, the neoplasm was completely excised and a computed tomography (CT) scan with contrast of the chest was performed, which showed a 3mm right upper lobe pulmonary nodule. However, a repeat CT scan of the patient was performed six months after excision that showed the pulmonary nodule had decreased in size to 1.5mm.



Figures 1A and 1B. A solitary violaceous plaque on the right flank.

Discussion

Cellular neurothekeoma (CNTK) is an uncommon benign cutaneous neoplasm of uncertain lineage that most commonly occurs on the head, neck, or upper extremities of young female patients usually less than 30 years of age. Clinically, the lesions are usually asymptomatic, slow-growing dermal nodules less than 2cm in diameter that may clinically resemble a cyst or dermatofibroma. A rare case of agminated lesions has been described.[1–3]

The neoplasm is usually centered in the dermis with involvement of the upper subcutis in some cases and is composed of multiple nodules of epithelioid cells with pale eosinophilic cytoplasm. Occasional mild nuclear atypia and a low mitotic rate are seen in a majority of cases. CNTKs are distinguished from myxoid neurothekeomas (also known as nerve sheath myxomas) by the absence of immunoreactivity for S-100, the less prominent spindled component and the relative lack of myxoid matrix in the former.[2] Immunohistochemical staining is negative for epithelial, melanocytic, and muscle markers and is usually positive for relatively nonspecific markers, such as NK1/C3, S100A6, CD10, vimentin, and PGP9.5.[1,2] Although it has been suggested CNTKs are of neural origin, other evidence points to myofibroblastic origin due to occasional positivity for smooth muscle actin and recent results from ribonucleic acid microarray experiments.[1,4]

Histologically, CNTKs may be confused with other entities and can pose a diagnostic problem for pathologists by mimicking melanocytic neoplasms, pilar leiyomyomas, superficial cutaneous myxomas, and PFHTs. Further complicating matters is that CNTKs occasionally have atypical features, such as larger size, extension to muscle or subcutaneous fat, increased number of mitoses, and pleomorphism. Despite these atypical features CNTKs still display benign behavior.[5]

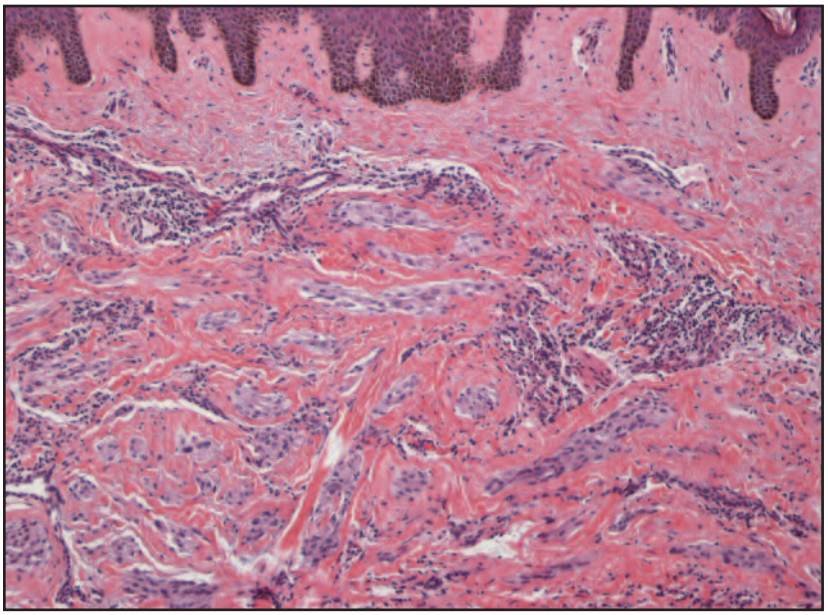


Figure 2. Nodules of epithelioid cells with pale cytoplasm and mild nuclear pleomorphism in the dermis.

The plexiform morphology of CNTKs can mimic many other dermal lesions that exhibit a plexiform pattern.[6] Immunohistochemical stains distinguish CNTKs from many of these plexiform lesions including myxoid neurothekeomas, Spitz nevi, and pilar leiyomyomas, but not always from PFHTs. Both CNTKs and PFHTs share similar demographics, cytology, and immunohistochemical staining and at times have overlapping histological findings.[7] Traditionally, CNTKs were thought to be dermal and PFHTs subcutaneous. Some authors believe that the two tumors may be related with PFHT representing the deeper subcutaneous variant. However, dermal PFHTs and subcutaneous CNTKs have been shown to exist, further obscuring the distinction.[7,8]

The distinction between the two entities is important as CNTKs are benign neoplasms that may occasionally recur while PFHTs are considered borderline neoplasms that have higher recurrence rates with rare case reports of metastases to regional lymph nodes or to the lungs.[8,9] This may be a reflection of the frequently deeper location of PFHTs compared to CNTKs.[8,9] Ultimately, if the lesion is not clearly a CNTK and there is a deeper component suggestive of a PFHT, complete excision and imaging may be warranted.

References

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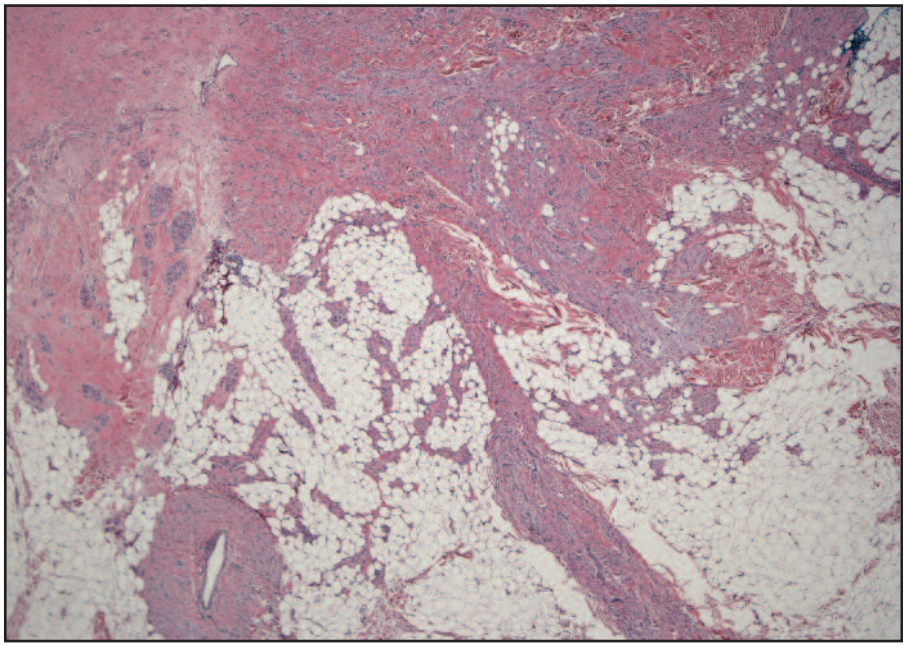

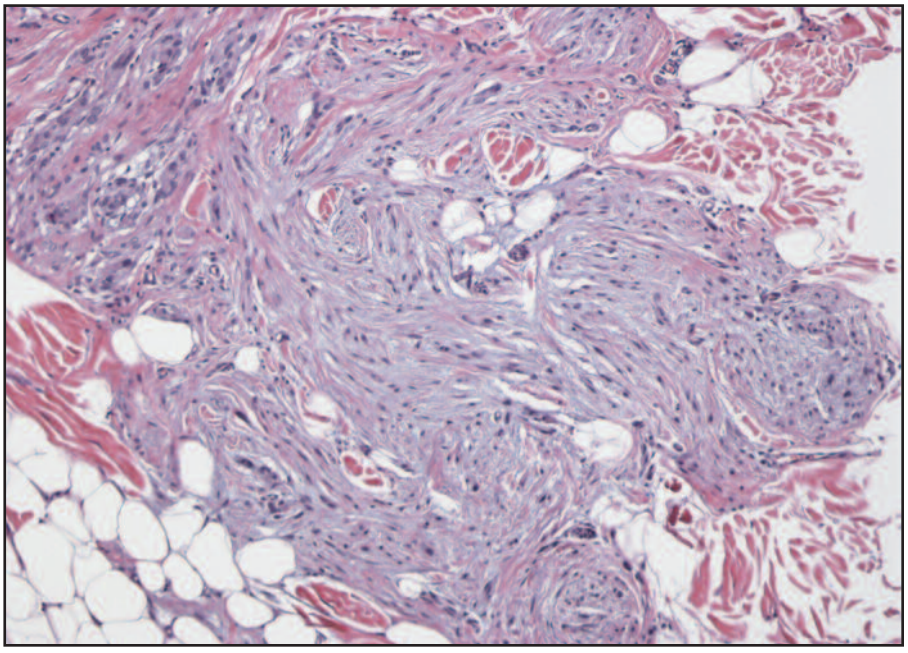

Figures 3A and 3B. Spindled component with a marked plexiform growth pattern deeper in the subcutis with features resembling a plexiform fibrohistiocytic tumor

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